IS THERE A PRIMARY CORD LESION IN PSEUDO-HYPERTROPHIC PARALYSIS?

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THIS is not a paper presented to set forth all that is new or interesting on this subject, but is merely the record of a case to supplement the appearances of some microscopic specimens Dr. Amidon has kindly made for me. Neurological medicine still possesses charms for me, but my work for the past two years has been confined to a different branch, yet a branch intimately associated with the one that calls us together on this occasion. I must, therefore, ask indulgence if I fail to have informed myself on the results of important investigations. The record of cases completed by autopsy, and an autopsy "worked up" by one so well known in neurology as the gentleman who shares this report with me, cannot fail to elicit interest and to excite discussion.

Professor Charcot, in a communication to *Le Progrès médical*, No. 10, 1885, and his pupils, Marie and Gerinon, in the *Revue de Médecine* for October, 1885, have classified the primary muscular atrophies, and in an editorial in the *Medical News* for December 19, 1886, this classification is discussed.

Until this had come under my notice I confess that I entertained the belief that the lesion in pseudo-hypertrophic paralysis was more frequently located in the spinal cord than these distinguished authors seem to think.

They recognize two distinct groups of cases: the spinal, and the primary myopathies: the lesions of the former being in the spinal cord, while the lesions of the latter are in the

muscles themselves. One includes all those pathological processes which involve the gray matter of the cord directly or indirectly. Amyotrophic lateral sclerosis and chronic polyomyelitis anterior are in the one group, while the three following are included in the latter:

Pseudo-hypertrophic paralysis, the juvenile form of progressive muscular atrophy of Erb, and the hereditary infantile form of paralysis of Duchenne.

Charcot's school endeavors, and the writer of the editorial above mentioned thinks quite successfully, to prove that these diseases are one and the same, differing in clinical features at some time during their course, and they should, therefore, be designated as *myopathie progressive primitive.* The lesion is thus regarded as an interstitial myositis, there being a variable production of adipose tissue and occasionally an hypertrophy of individual muscular fibres.

For many years I have been able to distinguish quite readily between a progressive muscular atrophy and a pseudo-hypertrophic paralysis. The clinical features are sufficiently differentiated in the text-books. The later stages are strikingly similar in the completeness of the atrophy and in the consequent helplessness of the victims. What surprises me is that the spinal cord has not been found diseased.

The case on which this paper is based came under my observation April 10, 1878, remained under my daily observation for one year, lacking two days, was reported to me from time to time subsequently, and terminated in death from catarrhal bronchitis December 30, 1885.

The patient was a male eight years of age when he first came under my care in the hospital. The loss of power dates from the time he began walking—the second year of life. The mother did not become alarmed, however, and did not seek medical advice, until 1875, his sixth year—now ten years ago.

The family is decidedly neurotic. The mother and all the children I have seen—four or five in number—have some peculiarity of speech. Three children have died of tubercular meningitis, so reported. One brother, two years

younger than the subject of this history, is affected in the same manner, and his case is progressing the same way. He had at one time large calves, but when he ceased walking the hypertrophy began to fade, and now the muscles are flabby and small, like those in other parts of the body.

When Dr. Amidon told me that he had partially examined the specimens I understood him to say that the findings would show marked and undeniable pathological changes.

His final report, however, leaves me still in doubt as to whether the changes are primary or secondary.

DR. AMIDON'S REPORT.

Cord examined from the filum to the decussation of the pyramids.

The only lesion appeared to be in the ganglion cells of the anterior horns. As compared with corresponding sections from a healthy cord, about one half the cells seem to have disappeared—leaving no trace. Those that remain are most of them poorly defined, small, and in many instances processless. This lesion is more marked in the dorsal and lumbar regions. The cells of the columns of Clarke seemed normal.